Case Report

Right ventricular thrombus with pulmonary artery aneurysm in a young male: A rare presentation of Behçet's disease

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ABSTRACT

We describe an adolescent patient presenting with hemoptysis. Detailed clinical work up of the patient showed right ventricular thrombus and bilateral pulmonary artery aneurysms along with the prescribed criteria for the diagnosis of Behcet's disease. Younger age of the patient was another distinctive feature of this case. Six months of therapy with cyclophosphamide and prednisolone resulted in near complete clinicoradiological response.

KEY WORDS: Behcet's disease, intracardiac thrombus, pulmonary artery aneurysms

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INTRODUCTION

Behcet's disease (BD) is a multisystem, chronic inflammatory disorder of unknown etiology. First described by Behcet Hulusi in the year 1937 in a patient with distinct picture of oral and genital ulcer with iridocyclitis.[1] Other organ involvement includes skin, joint, central nervous system, gastrointestinal tract, lung and cardiovascular system. This disease is frequent among the Mediterranean, Middle east and Far eastern population.[2]

Cardiopulmonary involvements include wide spectrum of abnormalities. Cardiac manifestations in Behcet's disease occur in about 1-5% of cases.[3] It includes coronary artery disease, recurrent pericarditis, myocardiopathy, and endocardiac abnormalities. Intracardiac thrombus formation is a rare and serious complication. It often occurs in association with pulmonary artery aneurysm (PAA). Young males seem to be most at risk and the right heart is the most frequent site of involvement.[4]

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Pulmonary involvement includes abnormalities of the vessel lumen and its wall, lung parenchyma, pleura and mediastinal structures.[5] PAA is reported in 1% of adults with BD. Pulmonary arteries are the second most common site of arterial involvement preceded by the aorta.[6] Aneurysms are more common than thrombosis.[6] Thrombosis of the pulmonary arteries is usually *in situ*.[7]

Considering the rarity of this disease and its distinct presentation with intracardiac thrombus and pulmonary artery aneurysm, we report this patient.

CASE REPORT

A 17-year-old male presented with hemoptysis of 3 days duration. The patient also complained of low grade fever, exertional dyspnea, decreased appetite and weight loss (7 kg) since last 6 months. Two and a half months back, patient noted pain and redness of both eyes and skin rashes on both thighs which resolved with some form of local treatment in 15 days. Further, on active questioning, the patient revealed recurrent oral and genital ulcers during last 1 year. The patient denied any history of similar illness in family.

General physical examination showed respiratory rate 16 breaths/minute, blood pressure 112/76 mm Hg, and pulse rate 80 beats/minute. Pallor, cyanosis, clubbing and lymphadenopathy were absent.

Examination of oral cavity [Figure 1] showed multiple oral ulcers. Scars of old ulcers were present on the scrotum.

Respiratory system examination revealed bilateral vesicular breath sounds. There were no added breath sounds including bruit. Ophthalmic examination was also normal.

Routine laboratory tests revealed hemoglobin 11.7 gm/dl, total leukocyte counts 9800/µl with neutrophils 75%, lymphocytes 20%, monocytes 3%, eosinophils 2% and ESR 100 mm in first hour. His fasting blood sugar, renal and hepatic functions were within normal limits. Serum immunological profile e.g. cANCA, pANCA, rheumatoid factor, antinuclear antibody and antibody to HIV were negative. Pathergy test was also negative.

Chest radiograph [Figure 2] showed a rounded poorly defined opacity of approximately 40 × 30 mm size in relation to descending branch of the right pulmonary artery. Rest of lung parenchyma and mediastinum were normal.

Contrast-enhanced computed tomography thorax [Figure 3] showed dilation of both pulmonary arteries containing thrombi. Walls of pulmonary arteries also appeared thickened. Lung window [Figure 4] showed focal area of pleural-based ground glass opacities/consolidation suggestive of pulmonary infarction in both basal lung fields.

CT angiogram thorax [Figure 5] was performed to delineate the details of pulmonary arterial anatomy. It showed dilatation with hypo dense soft tissue filling of the lumen of second order branch of the right descending pulmonary artery and third order branch of the left pulmonary artery. A soft tissue density focus in the right ventricle suggestive of thrombus was also present.

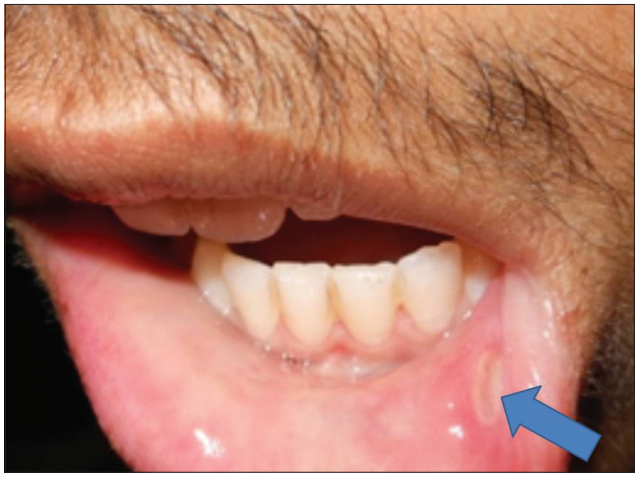

Figure 1: Photograph showing a shallow ulcer of about 12 mm × 4 mm size with raised and indurated margins over mucosal surface of the left lower lip

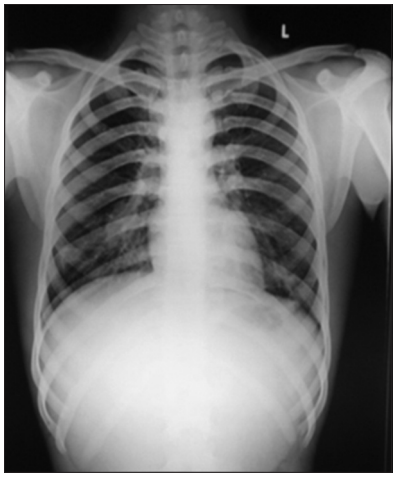

Figure 2: Chest X-ray PA view showing a poorly defined rounded opacity in relation to descending branch of the right pulmonary artery

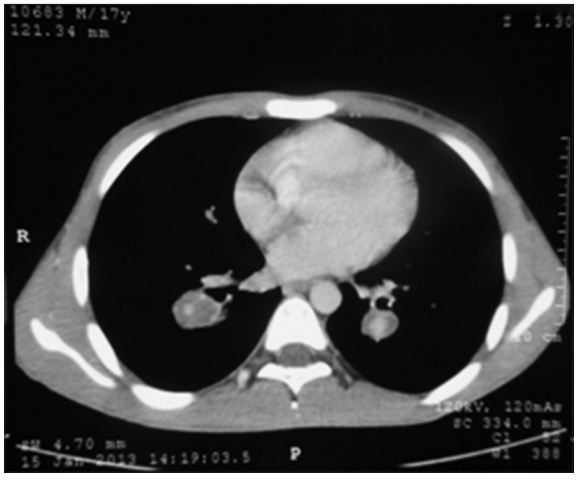

Figure 3: Contrast-enhanced computerized tomography of thorax showing an aneurysmal dilatation and thrombus in the bilateral descendant branches of pulmonary arteries

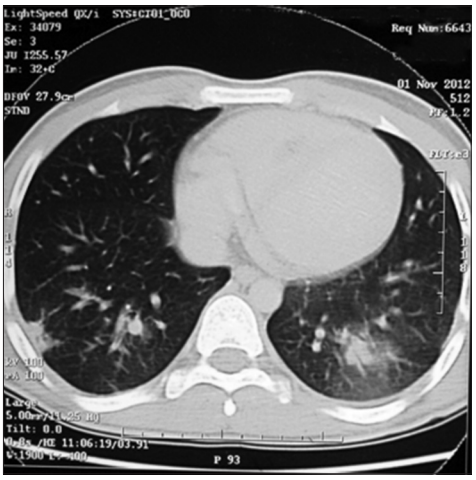

Figure 4: Multiple pleural-based ground glass opacities/consolidation in basal part of both lung fields

Echocardiography showed a 20 × 15 mm mass in right ventricular cavity with no evidence of any structural heart disease. Color Doppler of lower limbs showed normal venous flow.

In our patient, oral ulcer, genital lesions, vascular manifestations (PAA), skin, eye lesions and a negative pathergy test were present during disease duration. A systemic severity score of 7 out of 8 were present in our patient. Positive pathergy test is not necessary in all cases of Behçet's disease.[8,9]

The diagnosis of Behcet's disease with intracardiac thrombus and pulmonary artery aneurysm was made on the clinical and radiological findings.

The patient was given cyclophosphamide 1 gm along with mesna 600 mg intravenous monthly pulse therapy and tablet prednisolone 30 mg/daily tapered over 1 month to 10 mg daily. After 6 months of therapy, oral and scrotal lesions showed improvement. No fresh skin or eye lesion appeared. Repeat CT pulmonary angiogram showed near normal pulmonary arteries with partial resolution of right ventricular thrombus [Figure 6].

DISCUSSION

Present day diagnosis of BD is considered on the basis of newer international criteria for Behçet's disease (ICBD).[10] This new criteria has additionally incorporated vascular manifestations (VMs) e.g., superficial phlebitis, deep vein thrombosis, large vein thrombosis, arterial thrombosis and aneurysm to the earlier five criteria of international study group.[1]

Intracardiac thrombi in BD may result from endomyocardial fibrosis, which may be a sequele of vasculitis involving endocardium, myocardium or both.[11] As intracardiac thrombus is tightly attached to the endocardium, embolism from the cardiac cavity seems to be relatively uncommon.[12]

Young males seem to be most at risk than female and the right heart is the most frequent site of involvement for intracardiac thrombus formation.[4,13]

The diagnosis of intracardiac thrombi in BD may be made using cardiac magnetic resonance imaging, computed tomography, and transthoracic echocardiography, which may show a mass in the heart chambers, sometimes indistinguishable from infective vegetations or from a tumor and myxoma.[12,14]

In a series of 137 patients with BD only one patient was found to have right ventricular thrombus.[15] Similarly, only 1 patient was observed to have intracardiac thrombus out of 56 (1.78%) patients by Uçan *et al*.[16]

Recently, two BD patients with intracardiac thrombi and pulmonary artery aneurysms have been reported.[17,18] Luo *et al*.[19] analyzed the clinical characteristics of BD with intracardiac thrombus, diagnosed over a period of 1 year.

Pulmonary artery aneurysm (PAA) although rare is more commonly seen than intracardiac thrombi in patient of BD. Various other conditions which can also cause pulmonary artery aneurysm include trauma (often iatrogenic),[20] infections,[21] pulmonary hypertension,[22] congenital heart disease[23] and neoplasm.[24] A detail history and work up of patients is often sufficient to arrive an etiological diagnosis in a given patient. Helical CT is considered superior to MRI imaging in diagnosis of PAA.[6]

In a series of 534 patients with BD, only eight suffered from PAA and six of those died despite immunosuppressive treatment or surgery, underlining

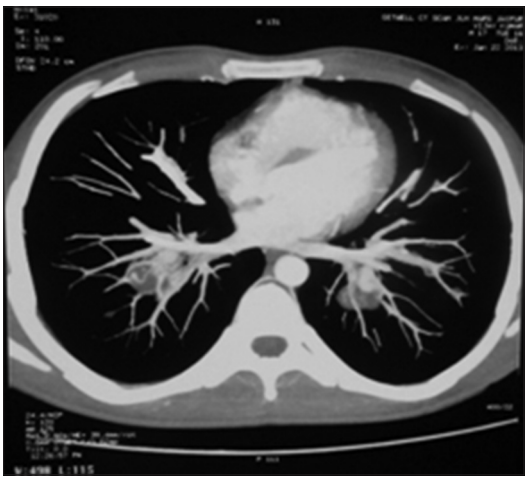

Figure 5: CT angiography at the level of pulmonary veins showing bilateral descending pulmonary artery aneurysms with thrombus in it. A small filling defect in the right ventricle suggestive of thrombus is also seen

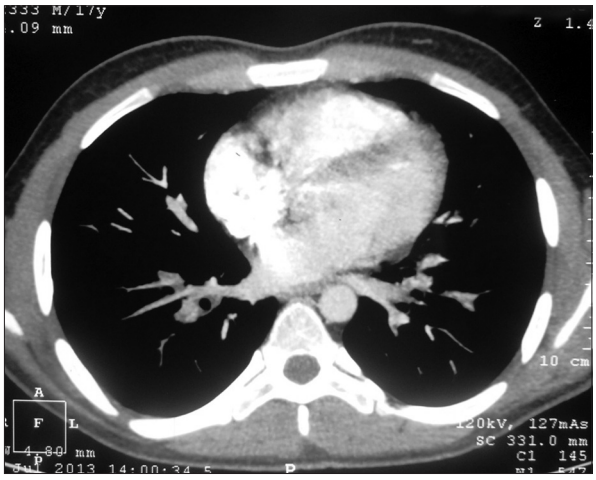

Figure 6: CT angiography of the thorax after 6 months of treatment showing marked reduction in the size of aneurysm of both pulmonary arteries. Thrombus in the right ventricle also appears smaller

the mortality associated with PAA.[1] Hamuryudan *et al*.[25] reported that 12 of 24 patients (50%) of PAA died after an average of 10 months after the onset of hemoptysis. However, Emad *et al*.[26] observed a higher frequency of PAA in 9 out of 16 patients with BD using multislice CT. Such higher frequency of PAA can be attributed to the excellent delineation of vessels lumen and wall on multislice CT scan.

Cyclophosphamide and corticosteroids are the main stay of medical treatment of this life-threatening disease carrying poor prognosis. However, early diagnosis and aggressive therapy result in remission with significant reduction in size of the pulmonary artery aneurysm and partial resolution of right ventricular thrombus. Surgical resection may be considered in patient with massive hemoptysis as a life-saving strategy. The major complication following surgery includes recurrence, false aneurysm and A-V fistula at the site of anastomosis. There are no controlled trial of anticoagulant and thrombolytic agents in BD.[15] Recently, Piga *et al*.[27] successfully used thrombolytic therapy for recurrent right ventricular thrombosis in a patient with BD. Due to the presence of hemoptysis in our case, we did not consider thrombolytic therapy.

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